Vascular Malformations

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KEYWORDS

- Vascular malformation • Capillary malformation
- Venous malformation • Lymphatic malformation
- Arteriovenous malformation • Glomuvenous malformations

Cutaneous vascular malformations are uncommon birthmarks that represent errors in vascular development and occur in approximately 0.3% to 0.5% of the population.[1] These lesions are much less common than infantile hemangiomas but are frequently confused with them. It is essential to properly diagnose these lesions because of their differences in morbidity, prognosis, and treatment.

CLASSIFICATION OF VASCULAR LESIONS

The classification of vascular anomalies has been hampered by the use of inaccurate terminology. Early classifications published by Virchow[2] characterized vascular lesions according to the vessel's pathologic appearance, dividing them into angiomas and lymphangiomas. The biologic behavior and natural history of the vascular lesions were not considered when classifying them. Thus, there was a tendency to identify any type of vascular anomaly as a hemangioma.

Mulliken and Glowacki[3] made great strides in clarifying this confusion when they published their landmark classification of vascular birthmarks in 1982, which grouped them into 2 major categories: hemangiomas and malformations. This classification has served as the foundation for the proper identification, investigation, and management of vascular birthmarks. Mulliken's biologic classification was modified slightly in 1996 to reflect new knowledge and the importance of other distinct types of vascular tumors, including the tumors that can cause Kasabach-Merritt phenomenon and others. The newer classification divided vascular birthmarks into *vascular tumors* and *vascular malformations*.[4] This classification of vascular anomalies has been widely

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adopted by clinicians and is the accepted classification of the International Society for the Study of Vascular Anomalies(ISSVA) (**Table 1**).

Although there are several types of vascular tumors, hemangiomas represent the overwhelming number of vascular tumors encountered by pediatricians. Hemangiomas are differentiated from vascular malformations by their clinical appearance, histopathologic features, and biologic behavior. Pediatricians should be most knowledgeable about differentiating hemangiomas from vascular malformations based on their clinical presentation. Hemangiomas are found to be more common in girls , whereas vascular malformations have an equal sex distribution. The natural course of hemangiomas involves rapid proliferation for the first several months of life with subsequent spontaneous regression, often leaving fibrofatty deposition, overlying anetoderma, and telangiectasias. Vascular malformations are often recognized at birth and grow proportionately with the child, with many becoming more prominent at puberty. In challenging cases, histopathologic evaluation, including immunohistochemical markers as well as radiologic studies, can further help to distinguish these two types of vascular anomalies (**Table 2**).[5–8] Despite these differences, the use of confusing nomenclature persists in the literature.

Vascular malformations can be further subdivided into groups based on vessel type and flow characteristics. Capillary, venous, and lymphatic malformations (LM) are slow-flow lesions, and arteriovenous malformations (AVM) and fistulae are fast-flow lesions. Combined lesions may also occur (see **Table 1**). Each type of vascular malformation is discussed in this article.

CAPILLARY MALFORMATIONS

Capillary malformations (CM), including fading capillary stains and port-wine stains, are among the most common vascular malformations affecting the skin (**Fig. 1**). True CM (the non-fading type) occur in approximately 3 of 1000 infants, are present

Table 1
Vascular anomalies' ISSVA/Mulliken classification 1996

Vascular Tumors	Vascular Malformations
Infantile hemangioma	Simple
Congenital hemangioma (RICH, NICH)	Slow-flow
Pyogenic granuloma	Capillary (C) malformation
Tufted angioma	Lymphatic (L) malformation
Kaposiform hemangioendothelioma	Venous (V) malformation
Hemangiopericytoma	Glomuvenous malformation
	Fast-flow
	Arterial (A) malformation
	Combined
	Slow-flow
	LVM
	CLVM
	CVM
	Fast-flow
	AVM
	CM-AVM
	AVF

Abbreviations: AV, arteriovenous; F, fistula; M, malformation; NICH, non-involuting congenital hemangioma; RICH, rapidly involuting congenital hemangioma.